ESSENTIAL THROMBOCYTOPENIC PURPURA HÆMORRHAGICA

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A RESUME of the literature cannot help but impress one with the difference of opinion extant respecting this disease, morbus maculosus Werlhofii, or essential thrombocytopenic purpura hæmorrhagica. A brief historical review of the subject is essential to clearly set forth the present status of our knowledge of this very interesting condition.

About 150 years ago Werlhof described this condition, but did not clearly differentiate it from various other forms of purpura. Several observers have added interesting opinions but not until recent years and the writings of Brill, Krumbhaar, Frank, Seeliger and others, have we been able to clearly differentiate between essential thrombocytopenic purpura hæmorrhagica and the various other forms of purpura.

All purpuras have as common characteristics hæmorrhage into the skin, mucous membranes, stomach, bowels, urinary tract, central nervous system, etc., and all, if severe, are followed by a secondary anæmia. All too are most common in the first two decades of life.

Essential thrombocytopenic purpura hæmorrhagica however alone presents the complete chain; clinical history, symptomatology and laboratory findings, which combined can only be found in this condition, and their combination serves to definitely differentiate this form of purpura from all the others. The clinical history is, in most cases, negative until the first symptom of hæmorrhage from the nose, vagina, gums or other mucous membranes, becomes sufficiently severe to cause alarm. This is followed by a progressive secondary anæmia which usually causes medical advice to be sought.

So far the disease may be any one of the purpuric conditions and recourse must be had to the laboratory to definitely settle the diagnosis. In a case of essential thrombocytopenic purpura hæmorrhagica it will be found that the blood clots normally, as to time, but the resultant clot is soft and non-retractile. The bleeding time is prolonged up to 20-40 minutes, and sometimes more, and last, but not least, the blood platelets, normally between 200,000 and 300,000 per c.mm. tend to fall in numbers to a few thousand, or to be entirely absent.

These various points,—absence of causative factors, and symptomatology combined with the above definite blood picture, will enable us to rule out all other idiopathic diseases, accompanied by hæmorrhage from the skin or mucous membranes, and to declare a case to be definitely one of essential thrombocytopenic purpura hæmorrhagica.

The case having been definitely diagnosed, what is the treatment? There is no treatment known for this condition which gives as good results as splenectomy. Local applications, styptics, the use of sera, transfusion and the use of various drugs as well as x-rays have been found to be of little, if any use. Cases have a tendency to become arrested for a while but will later recur and each recurrence is likely to be more severe than the previous one. One must therefore be of an open mind and not conclude that a certain line of treatment has effected a cure.

In 1916, Kaznelson of Prague, advised splenectomy for this condition and pointed out that it was for essential thrombocytopenic purpura hæmorrhagica alone that he so advised. Later it was taken up elsewhere and to date forty-five cases have been collected and reported by Clopton. Of these forty-five cases twenty-seven are reported as well, fifteen as improved, one unimproved and two died. The result therefore would fully justify surgical treatment, as expectant treatment shows a very much higher death rate, with very few classed as cured or even improved.

Operative technique is simple, but is apt to be accompanied by profuse hæmorrhage, particularly when the spleen is adherent to the diaphragm, in which case the division and ligation of adhesions becomes a matter of extreme difficulty. One should always therefore be prepared to do a transfusion during the operation, if cir-

cumstances so require, or in extreme cases to abandon all efforts to remove the spleen.

However, in spite of profuse operative hæmorrhage, most observers remark that as soon as the splenic pedicle is ligated, and the spleen removed, the hæmorrhage will stop and post operative hæmorrhages are no more frequent than in ordinary routine abdominal surgery.

Very shortly after splenectomy the blood platelets, however much reduced, rapidly increase in numbers and reach a point frequently very much above the normal. Thus it will be found that in twelve, twenty-four, or thirty-six hours the platelet count will be upwards of 200,000, and may attain 800,000 or 900,000 during the course of a few days. This high count will gradually subside to approximately the normal, or even fall below that point. It has been found however that, even though a level is reached far below the normal, hæmorrhages do not return.

Case Report.—V. R., a girl, aged seven, was first seen the 4th September, 1925, with the complaint that for about three weeks she had had painful swellings in various parts which were bright red and later passed through the usual stages of a bruise. Her personal history was that she was an ordinary full term baby, breast fed for eleven months, walked at one year and talked at two years. She has had mumps, measles, pertussis and scarlet fever; all very mildly; and the past illness one year ago. She has always been energetic and healthy, except as above, and has never shown any tendency towards hæmorrhage.

Her family history is negative.

About three weeks before the 4th September, 1925, her mother noticed a "bruise" on the sole of the child's foot and the following day a similar one was seen on the calf of her leg. Other hæmorrhages rapidly appeared, at first painful and then becoming itchy, on different spots of her lower extremities. Her appetite, usually good, failed and the child became listless and tired easily.

This was her condition when first seen and, on examination, some twenty hæmorrhagic spots of various ages were found on her lower extremities ranging up to the size of a half dollar. Apart from chronically enlarged tonsils and adenoids and carious teeth, the general examination was negative, and the spleen was at no time palpable.

Treatment with calcium lactate, hæmostatic serum, rest, etc., was instituted and the next day there had been several larger hæmorrhages on the legs and body. On the 6th September the blood picture was normal, except the platelets which were 170,000. On the 7th September, as her condition was much worse and hæmorrhages, (as yet all cutaneous), had increased, it was decided to resort to transfusion and her father was used as a donor. This was followed by little reaction, but no improvement, and two days later the torso was one mass of hæmorrhages, almost confluent, and some as large as the hand. In addition she had others on her legs and arms and an urticaria, non-hæmorrhagic however, on her face. That day, the 9th September, she complained bitterly of abdominal pain, passed tarry stools and vomited blood. A platelet count this same day was 150,000 per c.mm.

As the general condition was considerably worse splenectomy was advised, but refused by the parents, and further hæmostatic serum was given without result.

On the 14th September the platelet count was 120,000 and red cells were 4,300,000 and, in spite of very considerable hæmorrhage, the hæmoglobin was 85 per cent. From the 9th September to the 17th the stools were frequent and were full of blood, and the vomitus was always blood streaked, and frequently contained clots of partially digested blood. On this day, the 17th September the urine contained so much blood that it was almost black, and the abdominal pain was very severe. The following day the platelet count was 75,000 at 9 a.m. and 40,000 in the evening with a red cell count of 2,700,000.

Splenectomy was done the following day, the 19th September under ether oxygen anæsthesia, with very little hæmorrhage and followed by little shock and no vomiting.

Owing to a misunderstanding no platelet count was done until the 23rd September when it was found to be 150,000, but from the time of operation there were no more hæmorrhages except one small one on the wrist two days later. On the 26th September the platelets numbered 800,000 and on the 13th October 500,000.

The general condition improved rapidly, the only setback being that the child developed a very acute nephritis, about one month after

operation, due to the fact that she was exposed to cold, damp weather while improperly dressed.

On the 12th December the platelets numbered 350,000 and red cells 5,500,000 with hæmoglobin 95 per cent. The child at this time had recovered her usual spirits and strength and was rapidly increasing in weight towards her normal, though the urine still shows a trace of albumin.

It will be noted that:

1st. Non-surgical treatment absolutely failed and the condition got worse.

2nd. With the exception of one small one, two days after operation, there were no hæmorrhages after the spleen had been removed.

3rd. Platelet count rose rapidly to 800,000 per c.mm. and three months later it was 350,000.

4th. That the general condition at present is, (in spite of a severe nephritis), almost restored to normal.

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PYÆMIA AFTER SCARLET FEVER

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THE following case is an instance of a class which has been common in the past in any hospital treating a large number of scarlet fever patients, but which with more modern treatment should become exceedingly rare.

Doreen H. was a healthy little girl of five years, of healthy parentage. She had never been previously ill but contracted measles, the rash developing on April 21st, 1925. On April 23rd when convalescent she was accidently exposed to infection from scarlet fever. Three days later, April 26th, occurred the typical onset of scarlet fever with vomiting, headache, sore throat and a sharp rise of temperature to 102°. Within twelve hours the characteristic eruption appeared. For the first three days the disease appeared to be running a moderate course but on April 29th with a further rise of temperature to 104° the disease took on the characteristic septic type with profuse purulent discharge from the nostrils and mouth and on the following day a discharge from both ears. Now there were two occasions when this sequence might have been prevented. On exposure to infection she might have been immunized by a small dose of scarlet fever antitoxin or on developing the disease its course might have been aborted by a moderate treatment dose of the serum. However, at that time the serum was difficult to obtain and was only used in the worst cases, and her case appeared at first to be of only moderate severity. To resume, following the discharge from the ears her condition appeared to improve and the temperature to moderate but on May 7th with a further rise of temperature there was evidence of involvement first of the right and then of the left mastoid, necessitating a bilateral mastoidectomy on May 9th. Her fever did not subside after the operation and on May 12th there developed an acute arthritis of the right knee, followed by arthritis of the right ankle on May 14th. On the 16th both joints were aspirated showing thick pus with numerous hæmolytic streptococci. A blood culture taken the same day revealed a copious growth of the same streptococcus. Other joints became involved in rapid succession, the left sterno-clavicular, both elbows and both ankles.